



## SJÖGREN'S SYNDROME: Clinical Diagnosis & Management of Oral Complications

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### Purpose

Sjögren's syndrome is a systemic autoimmune disorder that features destruction of lacrimal and salivary gland tissues. Such destruction often results in keratoconjunctivitis sicca and xerostomia (dry eyes and dry mouth) (1,2). The dental officer often is the first healthcare practitioner to identify a patient with Sjögren's syndrome as the disease may initially present with oral signs and symptoms. Prompt diagnosis, treatment, and management of the oral and ocular sequelae are paramount to both patient and practitioner as 10 years or more may pass between the onset of signs and symptoms and the definitive diagnosis (3). This Clinical Update highlights the oral aspects of Sjögren's syndrome, offers current treatment guidelines, and stresses the need for long term comprehensive patient management.

### Etiology/Epidemiology

The term "autoimmune" defines a group of diseases that are caused by serum autoantibodies that react with a patient's own tissue. Sjögren's syndrome is of autoimmune origin and is characterized histologically by a lymphocytic infiltration of the major and minor salivary glands that destroys the normal acinar architecture. This produces a decrease in salivary and glandular secretions, resulting in oral and ocular dryness. Salivary and lacrimal dysfunction may be moderate to severe and, without treatment, is thought to be permanent.

Sjögren's syndrome is estimated to occur in over 1 million Americans (3,4,5). It has a strong predilection for women (80-90% of cases) usually in the 5<sup>th</sup> decade and is rare in children. Two distinct disease forms exist: **Primary Sjögren's syndrome** features salivary and lacrimal involvement (dry eyes/ dry mouth) with no associated connective tissue (CT) disorder. **Secondary Sjögren's syndrome** features salivary and/or lacrimal involvement with an associated CT disease. The most common CT diseases associated with Secondary Sjögren's syndrome are - rheumatoid arthritis, systemic lupus erythematosus, progressive systemic sclerosis (scleroderma), polymyositis, and primary biliary cirrhosis (2,4,6). Extraglandular involvement of the skin, nerves, lungs, kidneys, and vasculature may occur in both disorders (1) and may include lymphadenopathy, Raynaud's phenomenon, interstitial nephritis, interstitial lung fibrosis, vasculitis, and peripheral neuropathies (2,7). The most serious extraglandular complication of Primary Sjögren's syndrome is the significantly increased risk (40X) of developing lymphoma (2,3,6,7).

### Signs/Symptoms

Although variable signs and symptoms may present with Sjögren's syndrome, the principal oral symptom is xerostomia. Patients may relate complaints that include difficulty speaking, chewing and swallowing. These problems may be alleviated by drinking water. In addition, altered (dysgeusia) or diminished taste (hypogeusia), burning mouth complaints, and difficulty in wearing removable prosthetic appliances may be reported. Fatigue and depression are common (2,7,8). Clinical signs may include: diffuse, bilateral enlargement of the major salivary glands, a dry atrophic oral mucosa, candidiasis, fissured tongue with a loss of the filiform papillae, and in-

creased cervical and incisal caries (2,3,6,7,9,10). Decreased lacrimal flow may produce a scratchy/gritty feeling in the eyes that can induce corneal ulceration. Patients may report dryness in other sites such as the skin and nasal or vaginal mucosa. The following guidelines are offered when evaluating a patient for Sjögren's syndrome:

**1) Health questionnaire review:** A thorough review of the patient's current and previous medical history and chief complaint should be performed. Information should be sought regarding prescription and over-the-counter medications, a prior history of radiation treatment and any associated systemic conditions (4,6). In addition, a history of alcohol, caffeine, and tobacco usage should be obtained as these agents have a dehydrating effect on the oral tissues.

**2) Clinical examination:** A complete head and neck exam should include glandular and lymph node palpation to rule out enlargement, tenderness or masses. An assessment of the dryness of the oral mucosa can be performed by evaluating the presence or absence of saliva, angular cheilitis, active caries, a fissured and atrophic tongue, as well as epithelial debris or food in the vestibule. Dental instruments may stick to the oral mucosa and the saliva may be very thick.

**3) Initial assessment of salivary function:** Salivary glands should be examined and milked to determine saliva output. An objective evaluation of salivary flow rate (ml/min of whole saliva produced) can be obtained with a collection device called a Carlson-Crittenden collector - a small pipette-like device with suction. Resting saliva samples are collected first without stimulation, and then citrate is applied to the tongue to collect stimulated saliva. Salivary samples are collected for 5 minutes then analyzed for weight, total flow rate and chemistry. Though it is very difficult to determine what the normal range of salivary function is, some investigators believe an unstimulated whole salivary flow rate of < 0.1 ml/min and a stimulated flow rate of 0.5 ml/min is abnormal (3,4). More specialized diagnostic techniques include salivary gland imaging and a labial gland biopsy (11). Salivary gland imaging techniques such as scintigraphy, sialography, MRI or CT Scan may be utilized in the diagnostic workup.

The definitive diagnosis of SS often involves a labial lip biopsy and tissue examination of the minor salivary glands in the lower lip. The histo-pathology report will feature a chronic lymphocytic infiltration with noted destruction of the salivary acinar units.

**4) Serologic Evaluation:** Several laboratory tests are commonly ordered. The CBC with differential may demonstrate anemia. An elevated erythrocyte sedimentation rate, total immunoglobulins (IgG, IgM), total serum protein, a positive rheumatoid factor and antinuclear antibody (ANA), may be noted in addition to elevated levels of the autoantibodies anti-SS-A (anti-Ro), anti-SS-B (anti-La) (4,7).

**Diagnostic Criteria:** At present, no universally accepted criteria for diagnosis of Sjögren's syndrome exists. Since the cause of the condition remains unknown, no single test provides absolute evidence of the disease (3,5). All patients have lymphocytic infiltrates in their salivary and lacrimal glands (2). Decreased lacrimal flow produces characteristic corneal and conjunctival epithelial pathology that

can be revealed by staining with rose bengal dye and observed through a slit lamp. In addition, the reduced tear meniscus and break-up time, and Schirmer's tear test may provide vital diagnostic information. Diagnosis of primary Sjögren's syndrome usually requires the following three components: a minor salivary gland biopsy demonstrating focal lymphocytic infiltrates, evidence of lacrimal gland dysfunction, and serum autoimmune reactivity (+ autoantibodies, or elevated total serum immunoglobulins). Secondary Sjögren's syndrome requires the presence of an additional connective tissue (CT) disorder.

**Management:** Treatment of the patient with Sjögren's syndrome is palliative and supportive in nature. Saliva plays a critical role in maintaining the oral hard and soft tissues as well as providing lubrication and antimicrobial properties against bacteria, viruses and fungi. A decrease in saliva can result in increased caries, abrasion, erosion, candidiasis, and gingival inflammation. Wetting agents such as artificial saliva are usually formulated with a carboxymethylcellulose base (i.e., *Xerolube®* or *Salivart®*) and have met with varying degrees of acceptance. Salivary flow can be stimulated with sugarless candies such as lemon drops and sugarless gum. Pilocarpine hydrochloride (*Salagen® 5-10mg tid*) is a parasympathomimetic agent that can effectively stimulate salivary flow if functioning glandular tissue remains. Side effects include increased sweating, flushing, and hypertension (4,9). Pilocarpine is contraindicated in the presence of glaucoma, acute cardiac failure, bronchial asthma, urinary tract obstruction, peptic ulcer disease, hyperthyroidism, or Parkinson's disease (4,12). Medications with anti-cholinergic effects should be avoided as they can cause xerostomia.

**Preventive therapies:** Preventive dental care for oral soft and hard tissues is critical for all patients with Sjögren's syndrome. Daily application of a topical fluoride to remineralize enamel and prevent caries is the "cornerstone" of preventive treatment. Ideally, neutral sodium fluoride in a custom carrier is recommended to ensure adequate interproximal distribution and minimize tissue irritation (2,8,11,13). Antifungal therapy should be prescribed as needed. Nystatin oral suspension has a high sucrose content and may be problematic in the Sjögren's syndrome patient at high risk for caries. Systemic medications such as fluconazole (100 mg daily) or ketoconazole (200 mg daily) may be more advantageous.

**Nutritional/dietary analysis:** Patients should be encouraged to increase their water intake to at least 8 glasses per day. Dry, bulky, spicy, or acidic foods and those high in sugar should be avoided. Alcohol, caffeine, carbonated beverages, and alcohol containing mouth rinses should be avoided.

**Restorative:** Conservative restorations should be placed initially, until caries is under control. The margins of prostheses should be supragingival to facilitate cleaning and fluoride delivery. Amalgam is an excellent restorative material when restoring both cervical and posterior lesions and is more successful than composite materials (2,12). Where esthetics is of concern in anterior teeth, light cured glass ionomers with fluoride may be beneficial, however marginal integrity may be compromised in the dry environment.

**Recall Intervals:** Dental recall visits should be scheduled every 3 to 4 months for examination and prophylaxis (3,4,12,13). The importance of meticulous oral hygiene should be reinforced at each visit. Patients must understand their role as co-therapists in successful management of their condition (3).

## Conclusion

The diagnosis and treatment of Sjögren's syndrome involves a multi-disciplinary approach. Dentistry is an essential component in that approach. The goals of patient management include relief of symptoms, control of oral disease, improvement in residual salivary function, and long-term follow-up with an individualized preventive dentistry program (4,11,12).

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